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Case report

Therapeutic management for patients with cleft lip and palate complicated by sleep apnea syndrome: A case report

Tomoki Kato[a,b,*], Meiho Nakayama[c,d], Nagato Natsume[a,b]

[a] Division of Research and Treatment for Oral and Maxillofacial Congenital Anomalies, School of Dentistry, Cleft Lip and Palate Center, Aichi-Gakuin University, Nagoya, Japan
[b] Cleft Lip and Palate Center, Aichi-Gakuin University Hospital, Nagoya, Japan
[c] Good Sleep Center at Nagoya City University, Nagoya, Japan
[d] Department of Otorhinolaryngology at Nagoya City University, Nagoya, Japan

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ABSTRACT

We treated a patient with cleft lip and palate by palatoplasty, and he subsequently developed sleep apnea. We then performed tonsillectomy, to improve the sleep apnea, and were able to do this successfully without impairing velopharyngeal function. This successful result was achieved by collaboration between the oral and maxillofacial surgeons who initially performed the palatoplasty and the otolaryngologists who treated the patient for sleep apnea. The decision to remove the palatine tonsils was based on careful observation of the respiratory and sleeping conditions of the patient during sleep, together with polysomnography (PSG). The tonsillectomy was done using a two-step procedure. We obtained a good treatment outcome in a patient with cleft lip and palate who had sleep apnea, based on collaboration between otolaryngologists and oral and maxillofacial surgeons.

1. Introduction

During palatoplasty, the soft palate is pushed backwards to obtain good velopharyngeal function and accurate articulation [1–3], resulting in narrowing of the upper respiratory tract. In addition, after palatoplasty, the palatine and pharyngeal tonsils undergo physiologic enlargement in early childhood, resulting in more serious stricture of the tract. Both of these factors are causes of sleep apnea. If the bilateral palatine tonsils are extirpated to improve sleep apnea developing after palatoplasty, velopharyngeal insufficiency may result.

We treated a patient with cleft lip and palate by palatoplasty, and he subsequently developed sleep apnea. We then performed tonsillectomy, to improve the sleep apnea, and were able to do this successfully without impairing velopharyngeal function. This successful result was achieved by collaboration between the oral and maxillofacial surgeons who initially performed the palatoplasty and the otolaryngologists who treated the patient for sleep apnea. The decision to remove the palatine tonsils was based on careful observation of the respiratory and sleeping conditions of the patient during sleep, together with polysomnography (PSG). The tonsillectomy was done using a two-step procedure. Here, we describe the process leading to this good outcome, with reference to the literature.

2. Case report

The patient was a boy born by cesarean section at 38 weeks and 5 days gestation in a local obstetric clinic. He was the second child of a 33-year-old father and 30-year-old mother. Intrauterine growth retardation was identified, and he weighed 1965 g at birth. Because cleft lip and palate were identified at birth, he was referred to the pediatric department of a local hospital for whole-body examination. Staining of G-banded chromosomes performed there revealed a eupyrene 46, XY karyotype. At 28 days after birth, when he weighed 2436 g, the patient was referred to our center for treatment of the cleft lip and palate. Although cheiloplasty was initially scheduled to be performed when the child was 11 months old, the operation was postponed due to the sudden development of thrombocytopenia, diagnosed by the pediatric department of the local hospital. Cheiloplasty was rescheduled for when the boy was 12 months old, but again postponed due to chickenpox and thrombocytopenia. The child underwent cheiloplasty at 16 months of age, after the platelet count had normalized. Palatoplasty was

☆ AsianAOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

* Corresponding author at: Aichi-Gakuin University 2-11, Suemori, Chikusa-ku, Nagoya 464-8651, Japan. Tel.: +81 52 751 7181; fax: +81 52 759 2151.
E-mail address: t.kato.clpc@gmail.com (T. Kato).

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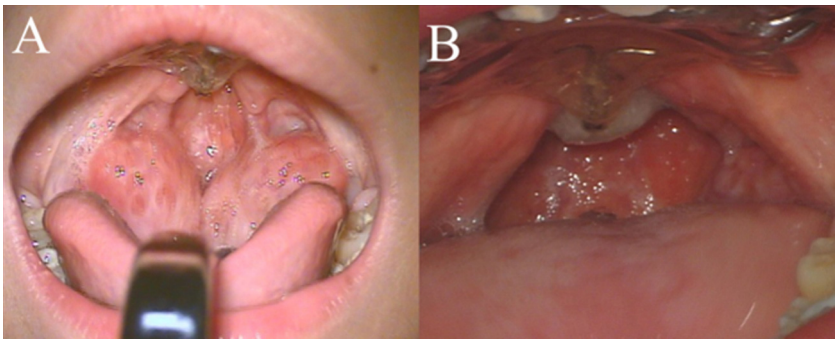

Fig. 1. Photographs of the patient's oral cavity. (A) The oral cavity before palatine tonsillectomy. The palatine tonsils are hypertrophied to the point that the tonsils on the left and right sides are nearly touching, like "kissing tonsils" central apnea. (B) The oral cavity after palatine tonsillectomy.

performed when he was 30 months old. Velopharyngeal insufficiency (VPI) occurred after palatoplasty. The VPI was corrected with a palatal lift prosthesis with attached bulb.

Recheiloplasty was performed when he was 8 years old, to reshape facial structures for esthetic reasons. An otolaryngologist at the local clinic had diagnosed the child with sleep apnea when he was 5 years old, but it had become progressively worse since then. Examination of the child at our center revealed that he sometimes fell into a state of somnolism. We therefore referred him to an otolaryngologist who is the chief director of the Good Sleep Center at Nagoya City University for closer examination (co-author Nakayama). Although the adenoid tonsils were not seriously hypertrophied, the palatine tonsils were hypertrophied to the extent that the tonsils on the left and right sides were nearly touching, like "kissing tonsils" (Fig. 1A).

The Good Sleep Center performed PSG to clarify the sleeping conditions. PSG uses the following investigations to clarify biophysiological changes: electroencephalography (EEG); electro-oculography; electrocardiography; chin electromyography, leg electromyography, nasal and oral airflow measurements, and respiratory plethysmography for movements of the chest wall and upper abdominal wall; and pulse oximetry for oxygen saturation. Polysomnographic technicians certified by the Japanese Society of Sleep Research performed the above studies overnight in a soundproof room at the Good Sleep Center while monitoring the patient with an infrared camera. Rechtschaffen and Kales scores [4] were used to evaluate the sleep stages and distinguish non-rapid eye movement (NREM) sleep from rapid eye movement (REM) sleep using EEG. NREM sleep comprises four stages. Stages 3 and 4 of NREM sleep are considered present when delta waves are apparent. Short-duration (2–15 s) and long-duration (>15 s) periods of arousal when the body was moving were recorded. The number of apneic events, total sleep time (TST), sleep efficiency (ratio of the number of minutes of sleep to the number of minutes in bed), apnea hypopnea index (AHI), and apnea index (AI) were also recorded. Obstructive apnea was defined as a cessation

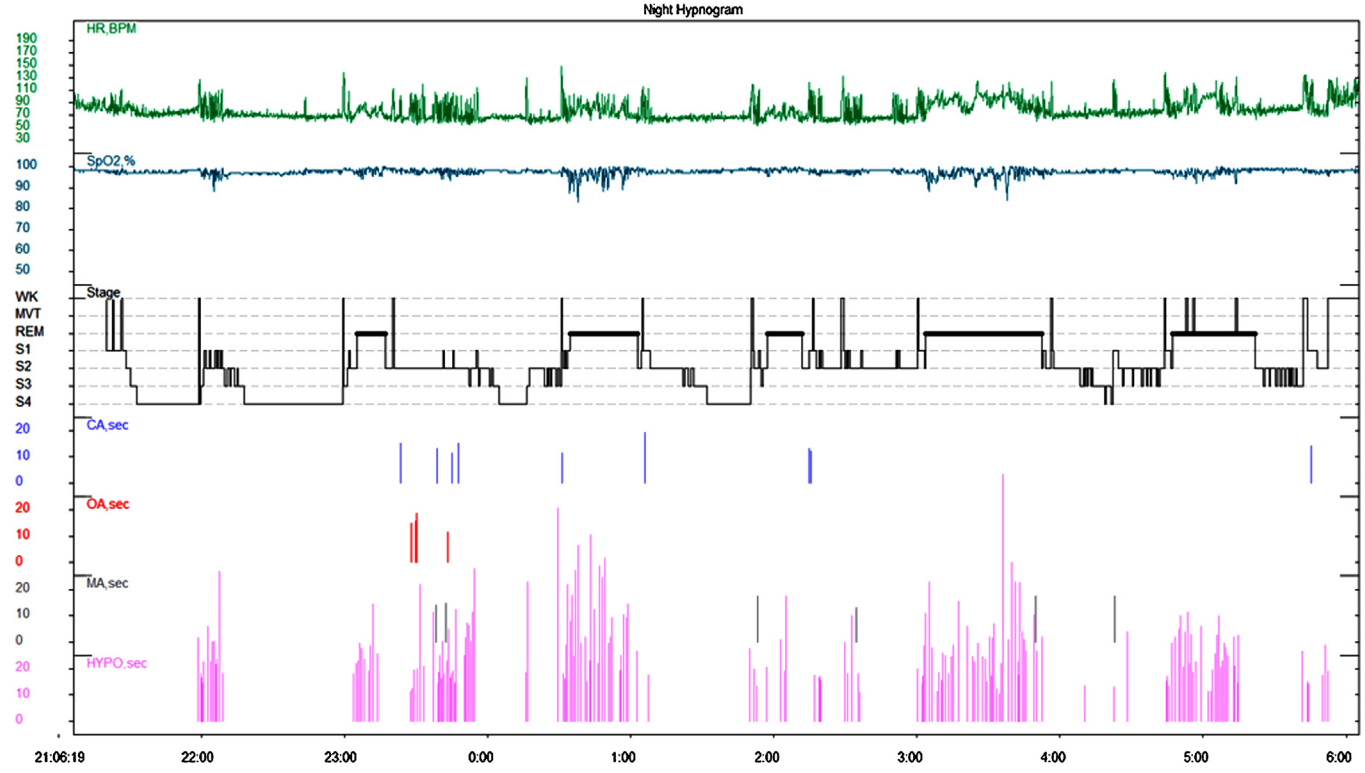

Fig. 2. Night hypnogram and respiratory findings before palatine tonsillectomy. HR BPM, heart rate beats per minute; SpO_2, arterial oxygen saturation; WK, wakefulness; MVT, movement; REM, rapid eye movement; S, stage; CA, central apnea; OA, obstructive apnea; MA, mixed apnea; HYPO, hypopnea; Sec, second.

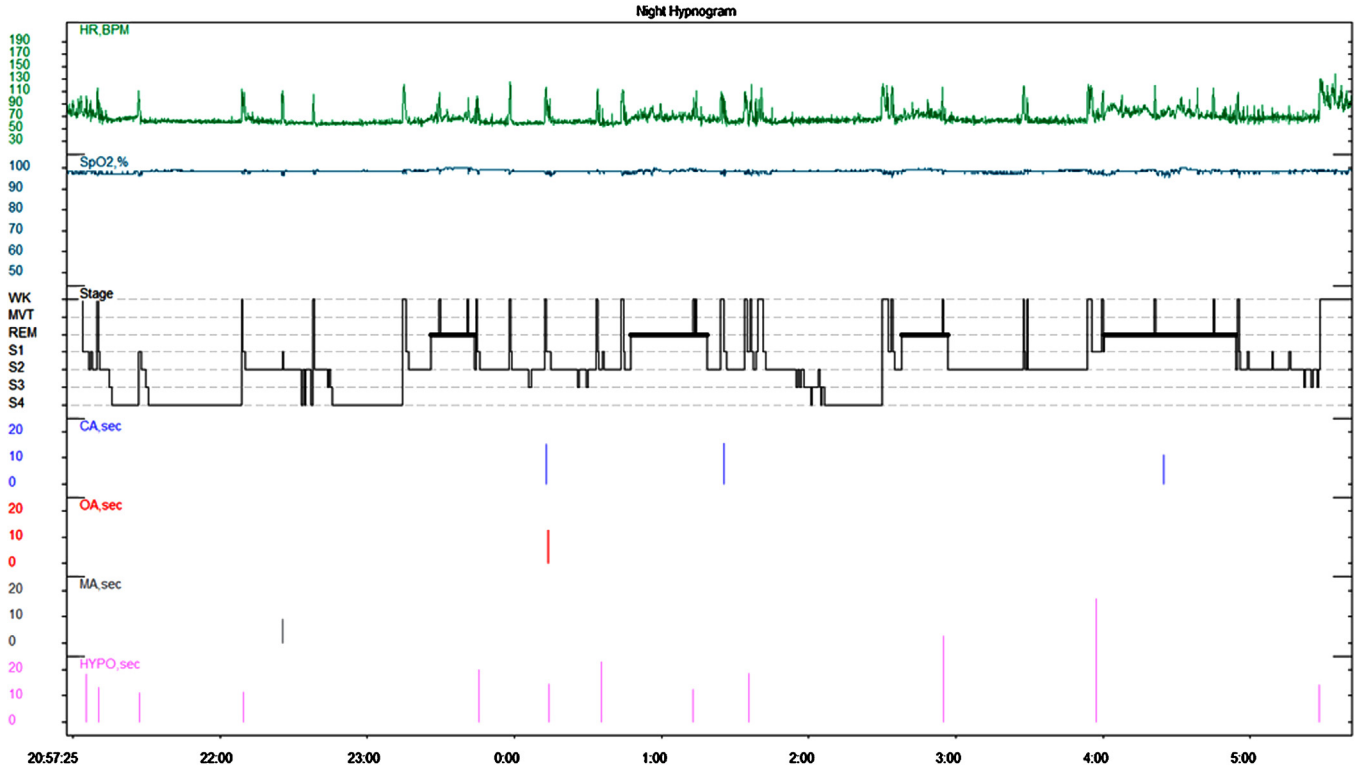

Fig. 3. Night hypnogram and respiratory findings after palatine tonsillectomy. HR BPM, heart rate beats per minute; SpO_2, arterial oxygen saturation; WK, wakefulness; MVT, movement; REM, rapid eye movement; S, stage; CA, central apnea; OA, obstructive apnea; MA, mixed apnea; HYPO, hypopnea; Sec, second.

of airflow through the nose and mouth for at least two respiratory cycles with paradoxical chest and abdominal movements. Hypopnea was defined as a >30% reduction in airflow through the nose and mouth with paradoxical respiratory effort resulting in either and arousal or an oxyhemoglobin desaturation of at least 4%. The obstructive apnea hypopnea index (AHI), defined as the average number of obstructive apneas and hypopneas per hour of sleep, was used for assessment of OSA severity. Apnea index (AI), hypopnea index (HI) and AHI were calculated by dividing the number of apneas or hypopneas per hour of sleep.

Before tonsillectomy, the patient had 19 apneic episodes (central apnea, 9; obstructive apnea, 4; mixed apnea, 6); AI, 2.3 events/TST hours; and AHI, 23.9 events/h during the night of observation (Fig. 2). In addition, average HR/BPM(heart rate beats per minute) was 97.6 during awake, 83.7 during REM stage, average SpO_2% was 97, while hypopnea (HYPO) revealed 181 times per TST.

Otolaryngologists usually extirpate the palatine tonsils bilaterally. Extirpation of the bilateral palatine tonsils, however, can lead to dysfunctional nasopharyngeal closure. We thought that extirpation of even one tonsil might improve the sleep apnea, and decided to perform palatine tonsillectomy on the right side first. The decision was comprehensively discussed with the patient's parents and informed consent was obtained (Fig. 1B). Our plan for the tonsillectomy was to first determine whether right-sided tonsillectomy could improve PSG scores without inducing dysfunctional nasopharyngeal closure, and if so, a second tonsillectomy would not be performed. Conversely, if the first tonsillectomy did not improve the PSG scores, the tonsil on the left side would be extirpated as well.

The PSG results after tonsillectomy of the right palatine tonsil revealed excellent improvement in the patient's sleep apnea: 5 apneic episodes (central apnea, 3; obstructive apnea, 1; mixed apnea, 1); AI, 0.6 events/TST hours; and AHI, 2.1 events/h, average HR/BPM was 94.6 during awake, 72.1 during REM stage, average SpO_2% was 98, while hypopnea (HYPO) revealed 12 times per TST (Fig. 3). In addition, a fiberscope test and hearing test performed by a speech pathologist revealed no deterioration of the functions of nasopharyngeal closure after tonsillectomy (Fig. 4).

3. Discussion

During palatoplasty the soft palate is pushed backwards to maintain velopharyngeal function [1–3], which results in narrowing of the upper respiratory tract. The pharyngeal tonsil enlarges during early childhood and reaches full size at the age of 3–5 years. The palatine tonsils also enlarge during early childhood, and reach full size by 5–7 years of age. This means that before around 5 years of age, the already narrowed upper respiratory tract of patients with cleft lip and palate who have undergone palatoplasty narrows further with growth of the tonsils. This contributes to an increased risk of sleep apnea in patients treated for cleft lip and palate. The peak of sleep apnea in children with enlarged adenoid and/or tonsils occurs from 2 to 8 years of age. Thus, close attention is needed in patients who undergo palatoplasty [5–8].

Palatine tonsillectomy or adenotonsillectomy is considered useful for improving sleep apnea in such patients.

Palatine tonsillectomy or adenotonsillectomy must be very carefully performed, however, because these procedures may impair the velopharyngeal functions that were maintained with great effort during palatoplasty. In addition, otolaryngologists generally take the approach of extirpating the bilateral palatine tonsils. Our team of otolaryngologists and oral surgeons attempted to devise the optimal treatment for this patient, i.e., treatment with palatine tonsillectomy or adenotonsillectomy, without adversely impacting the velopharyngeal function. To achieve this goal, the oral and maxillofacial surgeons who performed the palatoplasty maintained close communication with the otolaryngologists who performed the palatine tonsillectomy or adenotonsillectomy.

Palatine tonsillectomy is usually performed bilaterally, but tonsillectomy carries a risk of postoperative hemorrhage [9]. The

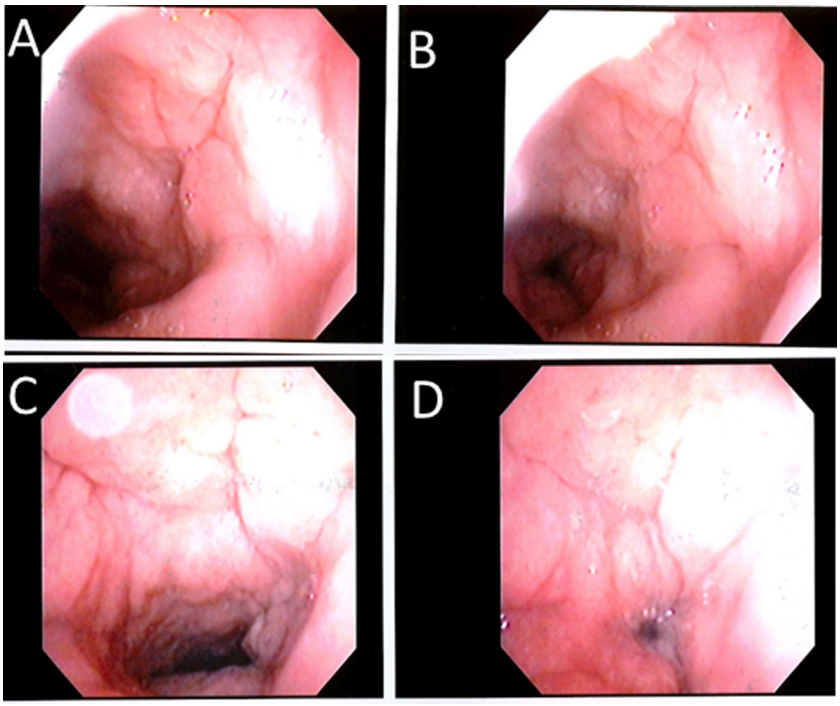

Fig. 4. Pictures of nasopharyngeal fiberscope. (A) Relaxed velopharynx before tonsillectomy. (B) Opening remains on voiced blowing before tonsillectomy. (C) Relaxed velopharynx after tonsillectomy. (D) Opening remains on voiced blowing after tonsillectomy.

mortality rate from post-tonsillectomy bleeding is reported to be 1 in 12,000 [10]. The American Academy of Pediatrics has stated that children with sleep apnea have a higher risk of hemorrhage in tonsillectomy [11]. If patients with sleep apnea undergo tonsillectomy on both sides at once, the rates of morbidity and mortality from post-tonsillectomy bleeding will presumably be higher [12–14]. In addition, removal of both tonsils may cause deterioration in the function of nasopharyngeal closure. To avoid these risks, we performed PSG before palatine tonsillectomy to precisely determine the respiratory and sleep conditions of the patient. Based on these findings, palatine tonsillectomy was performed on one side first. PSG was then performed again to determine how the first tonsillectomy improved sleep apnea, The results of this PSG were used to judge whether a second tonsillectomy should be performed. Adenotonsillectomy is the most common and first choice of treatment for children with OSA, as adenoidectomy alone may not be sufficient. In otherwise healthy children with adenotonsillar hypertrophy, polysomnographic resolution appears to occur in 75% or so after adenotonsillectomy. However, we hesitated to perform bilateral adenotonsillectomy at once because of its possible on VPI. Since it is well known that pediatric OSA can lead to morbidity, we decided to perform unilateral tonsillectomy after again obtaining informed consent from the patient's parents.

While pre-tonsillectomy PSG scores showed an AI of 2.3 events/TST hours and an AHI of 23.9 events/h, post-tonsillectomy PSG scores revealed definite improvements, with an AI of 0.6 events/TST hours and an AHI of 2.1 events/h. We therefore decided not to perform a second surgery. Furthermore, a speech-hearing therapist performed a fiberscope test and hearing test to ensure that the tonsillectomy did not impair the function of nasopharyngeal closure. These results indicate that, palatine tonsillectomy with PSG is effective for diagnosis and therapy in patients born with cleft lip and palate. Compared with conventional methods of tonsillectomy, our plan for tonsillectomy may cast a heavier short-term burden on patients and their families in that the patients may have to undergo PSG several times. Tonsillectomy in patients with sleep apnea, however, carries a higher risk of bleeding. Therefore, our tonsillectomy plan is safer, and thereby decreases the overall burden on the patients and their families. Because the number of centers specializing in sleep disorders is increasing, the use of reliable evidence-based procedures may minimize the surgical risks. At the same time, effective surgical methods should be developed in collaboration with these centers.

4. Conclusion

We obtained a good treatment outcome in a patient with cleft lip and palate who had sleep apnea, based on collaboration between otolaryngologists and oral and maxillofacial surgeons.

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